BIBLIOGRAPHIC INFORMATION SYSTEM

Journal Full Title: Journal of Biomedical Research & Environmental Sciences

Journal NLM Abbreviation: J Biomed Res Environ Sci

Journal Website Link: https://www.jelsciences.com

Journal ISSN: 2766-2276

Category: Multidisciplinary

Subject Areas: Medicine Group, Biology Group, General, Environmental Sciences

Topics Summation: 128

Issue Regularity: Monthly

Review Process type: Double Blind

Time to Publication: 7-14 Days

Indexing catalog: Visit here

Publication fee catalog: Visit here

DOI: 10.37871 (CrossRef)

Plagiarism detection software: iThenticate

Managing entity: USA

Language: English

Research work collecting capability: Worldwide

Organized by: SciRes Literature LLC

Manuscript should be submitted in Word Document (.doc or .docx) through **Online Submission** form or can be mailed to support@jelsciences.com

Vision

Journal of Biomedical Research & Environmental Sciences main aim is to enhance the importance of science and technology to the scientific community and also to provide an equal opportunity to seek and share ideas to all our researchers and scientists without any barriers to develop their career and helping in their development of discovering the world.

Journal of Biomedical Research & Environmental Sciences ISSN: 2766-2276
Subject Area(s): METABOLIC SYNDROMES | METABOLISM | PUBLIC HEALTH CASE REPORT

Lifestyle, Nutritional and Psychological Changes in Adult Patients with Inborn Errors of Metabolism during COVID 19 Pandemic: Results from an Online Survey

Gugelmo G[1], Schiff S[2], Lovato E[2], Lenzini L[3], Boscari F[4], Francini-Pesenti F[1], Avogaro A[4] and Vitturi N[4]*

[1]University of Padova, Department of Medicine-DIMED, Division of Clinical Nutrition, University Hospital, Padova, Italy
[2]University of Padova, Department of Medicine-DIMED, Clinica Medica 5, University Hospital, Padova, Italy
[3]University of Padova, Department of Medicine-DIMED, Emergency Unit, University Hospital, Padova, Italy
[4]University of Padova, Department of Medicine-DIMED, Division of Metabolic Diseases; University Hospital, Padova, Italy

ABSTRACT

When COVID-19 pandemic out broke in Italy, during the lockdown from March to May 2020, Inborn Errors of Metabolism (IEM) patients were at risk of not getting their dietary special products and routine visits. Moreover, during pandemic, psychological difficulties might have arose in these subjects, even more severe than in the general population due to the worries about acute decompensation caused by a possible COVID-19.

In order to evaluate the patients' perception of the outbreak situation and their related needs, three simple online anonymous surveys drawn up by Google Forms were sent to patients and families referring to our Adult IEM Center. Answers were collected between April and May 2020. Questionnaires investigated nutritional and lifestyle changes and psychological status using validated psychological tools.

19 patients with IEM filled out our survey (Median age 26-30 years). The most common nutritional therapy was low protein diet. During quarantine 12% patients failed to follow their usual medical diet, 65% reduced their physical activity and no one underwent an acute metabolic crisis. 57% of patients asked for more frequent access to the reference center. 33% of patients showed stress perceived of clinical relevance and general health perception were out of normal in 40% of patients.

In conclusion, during quarantine some patients reported difficulty in following their medical diet or physical activity and were clinically stressed. Despite this, no one experienced a metabolic crisis, but asked for contacting the Metabolic Team in different ways than usual due to worries about their health condition. Telemedicine, the possibility of clinical follow-up at home patient (Including blood tests) and reservation of non-COVID-19 beds for hospital admission of IEM patients may have contributed to help IEM adult patients in better face this emergency time.

INTRODUCTION

Inborn Errors of Metabolism (IEM) are rare disorders caused by genetic defects. In many of these disorders, metabolism of protein or fat or carbohydrates is affected. In order to achieve good metabolic control, treatment is often both pharmacological and nutritional. Children and adults affected by aminoacidopathies, organic acidaemias or urea cycle disorders require low protein diets to prevent excess

***Corresponding author**

Nicola Vitturi, Department of Medicine-DIMED, Division of Metabolic Diseases, University Hospital, Padova, Italy

Tel: +39-049-821-4326
E-mail: nicola.vitturi@aopd.veneto.it

DOI: 10.37871/jbres1197

Submitted: 13 February 2021

Accepted: 01 March 2021

Published: 02 March 2021

Subject: Medicine Group

Topic & Subtopic(s): Metabolic Syndromes; Metabolism; Public Health

VOLUME: 2 ISSUE: 3

How to cite this article: Gugelmo G, Schiff S, Lovato E, Lenzini L, Boscari F, Francini-Pesenti F, Avogaro A, Vitturi N. Lifestyle, Nutritional and Psychological Changes in Adult Patients with Inborn Errors of Metabolism during COVID 19 Pandemic: Results from an Online Survey. J Biomed Res Environ Sci. 2021 Mar 02; 2(3): 100-103. doi: 10.37871/jbres1197, Article ID: JBRES1197

accumulation of toxic metabolites. Only a part of natural foods can be allowed in their treatment, with consequent supplementation with special low protein foods and amino acids free of toxic precursor supplements [1]. Patients with disorders of glycogen storage and fatty acid oxidation often require carbohydrate supplements in order to maintain normoglycemia [1]. Furthermore, metabolic disorders affect catabolic processes (Breakdown of fatty acids, amino acids or glycogen), therefore promotion of anabolism and prevention of events such fever or infections are key elements for treatment [2]. Provision of emergency treatment during acute illness is essential for IEM at risk for decompensation. IEM patients and their families receive education to their specific nutritional therapies and to follow emergency regimens, through special products (Foods and drugs) prescribed by Metabolic Care Center. Moreover, patients and their families often need psychological support in order to better manage their lifelong therapy. In March 2020 COVID-19 outbroke in Italy and then worldwide, posing an unprecedented challenge for patients and the whole healthcare system. In particular during the national lockdown from March to May 2020, IEM patients had to face the risk of not to get their special products, not to have their usual management for routine visits and, at worst, in case of acute decompensation caused by a possible COVID-19 infection or even caused by other stress factors, of not following their usual emergency treatment. Furthermore, during pandemic, psychological difficulties might have arose in these subjects, even more severe than in the general population. In fact, patients and their caregivers might have been concerned for their routinely dietetic and pharmacological treatment and for the risk of decompensation for contracting a potentially dangerous virus. In order to give patients and clinicians better chances to overcome this pandemic, we asked our patients to tell us their nutritional and psychological changes during quarantine.

MATERIALS AND METHODS

Patients

Our center contacted 42 IEM patients and their families by email, asking them to fulfill two online surveys. 19 patients (12 males and 7 females) with rare metabolic diseases accepted to fill our questionnaires. The patients' median age was 26-30. Frequency of pathologies is described in figure 1. The most common nutritional therapy was low protein diet, mainly for Urea Cycle Disorders (UCD) and organic acidurias. None of the patients participating to the survey was affected by COVID-19 or had close contact with patients affected by Coronavirus.

All procedures followed were in accordance with the Helsinki Declaration of 1975.

Questionnaires

We elaborated two simple questionnaires (Supplementary material) that were sent by email to our patients and families. Answers were collected between April and May 2020. The questionnaires were anonymous and they were drawn up by Google Forms. The first survey was addressed to patients and caregivers both, and questions were about nutritional and lifestyle changes; the second questionnaire was redirected only to patients and its focus was to investigate their states of health and stress perceived by using validated psychological tools (Stress Scale Perceived (SSP) and General Health Questionnaire (GHQ_12)).

RESULTS

Questionnaire 1

Our Center drawn up a special questionnaire consisting of 29 multiple choice questions about: modification in diet

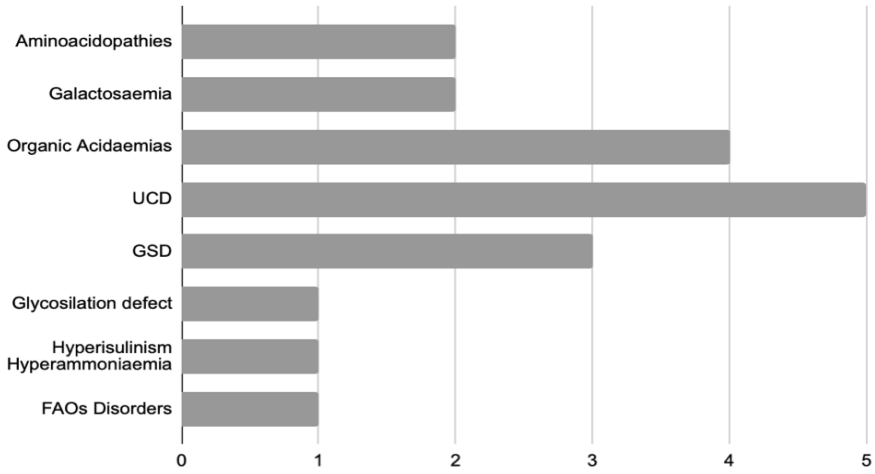

Figure 1 Frequency of IEM recovered by Questionnaires. Abbr. UCD: Urea Cycle Disorders; GSD: Glycogen Storage Diseases; FAOs: Fatty Acid Oxidation.

Table 1: Data from questionnaires.

Diet	
Modified meal time	29%
Increased quantity of food	18%
Failure to follow usual medical diet	12%
Augmented number of meals	12%
Difficulties in obtaining special medical food and drugs	6%
Physical Activity	
Reduced	65%
Enhanced	12%
Contact with Metabolic Team	
More frequent access to the reference center	47%
Request on information and advice about acute management of infection	29%
Request on information and advice about diet	23%
Request on information and advice about physical activity	18%
Preferred Communications Platform	
By email	47%
By web- conference	18%
By social network	18%
Psychological Status	
Stress perceived of clinical relevance	33%
General health perception out of normal	40%
General health perception near normal	33%
General health perception normal	27%

(Number and time of meals) and physical exercise; difficulty to obtain special medical food and drugs; needs of metabolic support during quarantine and how Metabolic Care Center may help them to overcome difficulties that could have been arisen; test positive or close contact with people affected by COVID-19.

During quarantine main dietetic and lifestyle changes reported by patients and caregivers are (Table 1): 12% of patients failed to follow their usual medical diet and only 6% declared difficulties in obtaining special medical food and drugs; 18% of patients increased the quantity of food, 29% modified the meal time and 12% augmented the number of meals, 65% reduced their physical activity, 12% enhanced it.

Despite the adjustments, only 17% patients gained weight, and no one underwent an acute metabolic crisis.

About the need of having closer contact from their Metabolic Team, 47% of patients asked for more frequent access to the reference center: 24% asked for information and advice about diet, 12% about physical activity, 29% about acute management of infection. 47% of patients declared preference of contact by email, 18% by web- conference and 18% by social network.

Questionnaire 2

Psychological questionnaires used for patients were two: Stress Scale Perceived (SSP), General Health Questionnaire (GHQ_12). We collected 15 answers by patients and results suggest these main psychological features (Table 1): 33% of patients showed stress perceived of clinical relevance (In SSP 5/15 subjects had z score > 2SD); the GHQ_12 (Referred to two weeks before the test has been fulfilled) revealed general health perception normal in 27% of patients (4/15), near to normal limits in 33% (5/15) and out of normal in 40% (6/15).

DISCUSSION

Information about the impact of COVID-19 on patients with IEM are currently very scarce. Case reports of IEM patients with confirmed viral infection have been published [3-5] but data about the patients' perception of the outbreak situation and their related needs are still lacking.

As indicator of the epidemic's impact on these patients, Elmonem, et al. [6] reported the effect of COVID-19 pandemic on the diagnosis and management of inborn errors of metabolism by considering the access to diagnostic and therapeutic services. This approach allowed obtaining useful quantitative estimates of health system effort but did not help to evaluate the effects on patients' day-life.

In fact, during the Italian national lockdown from March to May 2020, IEM patients had to face the risk of not being able to follow their usual medical diet or to access to health system services for their routine visits. As described by Brunetti-Pierri, et al. [7], the whole healthcare system and consequently also healthcare services targeted to IEM patients were reorganized: in our hospital all unnecessary visits had to be rescheduled and telemedicine (By email, videocalls, phone calls) became the major tool used to monitor IEM patients. Some beds non COVID-19 were reserved for possible admission of IEM patients or other chronical diseases.

Thus, to assess the main difficulties emerged during the Italian national lockdown, we conducted a survey among adult patients in our Metabolic Center focusing on their nutritional and psychological problems during the outbreak.

By this, we highlighted that during COVID-19 quarantine some adult IEM patients reported difficulty in following their medical diet or regular physical activity, due to inability to leave their house. Despite this, no one experienced a metabolic crisis, but frequently asked for contact with the metabolic team in different ways than usual.

Psychological questionnaires demonstrated general health perception (Referring to two previous weeks) out of normal in 40% of patients and stress perception was altered in 33% of patients. These alterations may be due to patients' concerns for a possible fatal infection and/or to the modification of day-life activities in fragile subjects with specific therapies and routines. Our data are consistent with a systematic review about mental health status in general population during COVID-19, in which Xiong, et al. [8] highlighted that prevalence of symptoms of adverse psychiatric outcomes among general population increased during pandemic period. Moreover, in that paper chronic diseases patients demonstrated more anxiety and stress symptoms. Risk factors identified for exhibiting these symptoms are poor economic status, lower education level, and unemployment [8], that are all possible risk factors in IEM adult patients.

This study has the limit of a selection bias: at first, although IEM are rare disorders, the sample of patients enrolled was small (Only 19 patients); in second instance, as adult IEM subjects are a new emerging population (Especially thanks to newborn screening set up in 1990s), patients of this study were relatively young, probably well-educated to use the internet, good healthy state and more positive mindset.

In conclusion, during quarantine some patients reported difficulty in following their medical diet or physical activity and were clinically stressed. Despite this, no one experienced a metabolic crisis, but asked for contacting the Metabolic Team in different ways than usual due to worries about their health condition. Although the forced reorganization of follow-up visits during lockdown period, some factors may have contributed to help IEM patients in better face this emergency time, such as alternative monitoring tools (i.e. Telemedicine), the possibility of clinical follow-up at home patient (Including blood tests) and reservation of non-COVID-19 beds for hospital admission of IEM patients.

DETAILS OF THE CONTRIBUTIONS OF INDIVIDUAL AUTHORS

NV and GG elaborated and analyzed questionnaires and wrote the manuscript; SS, EL and FB elaborated the questionnaires; FPF, LL and AA revised the manuscript.

ACKNOWLEDGEMENT

Authors have received research grants from Mamoxi/metaX S.r.L. Corso Francia 238-10146 Torino-Italy.

References

1. Dixon M, MacDonald A, White F, Stafford J. Disorders of amino acid metabolism, organic acidaemias and urea cycle disorders. In Shaw V, ed. Clinical Paediatrics Dietetics. 2015; Chichester: Wiley Blackwell. 381-525.
2. Van Hove JL. Anabolism: Practical Strategies. In Bernstein LE, Rohr F, Helm JR, eds. Nutrition Management of Inherited Metabolic Diseases. 2015; Cham: Springer International Publishing. 59-62.
3. Caciotti A, Procopio E, Pochiero F, Falliano S, Indolfi G, Donati MA, Ferri L, Guerrini R, Morrone A. SARS-CoV-2 infection in a patient with propionic acidemia. Orphanet J Rare Dis. 2020 Oct 28;15(1):306. doi: 10.1186/s13023-020-01563-w. PMID: 33115512; PMCID: PMC7592193.
4. Vitturi N, Lenzini L, Francini-Pesenti F, Gugelmo G, Avogaro A. SARS CoV2 infection in a young subject affected by arginosuccinate synthase deficiency: A case report of epilepsy worsening. Mol Genet Metab Rep. 2020 Dec 11;26:100698. doi: 10.1016/j.ymgmr.2020.100698. PMID: 33354515; PMCID: PMC7745051.
5. Lampe C, Dionisi-Vici C, Bellettato CM, Paneghetti L, van Lingen C, Bond S, Brown C, Finglas A, Francisco R, Sestini S, Heard JM, Scarpa M; MetabERN collaboration group. The impact of COVID-19 on rare metabolic patients and healthcare providers: results from two MetabERN surveys. Orphanet J Rare Dis. 2020 Dec 3;15(1):341. doi: 10.1186/s13023-020-01619-x. PMID: 33272301; PMCID: PMC7711270.
6. Elmonem MA, Belanger-Quintana A, Bordugo A, Boruah R, Cortès-Saladelafont E, Endrakanti M, Giraldo P, Grünert SC, Gupta N, Kabra M, Knerr I, Krämer J, Kuster A, Levtchenko E, Ngu LH, Rovira-Remisa MM, Sass JO, Sykut-Cegielska J, Tummolo A, van den Heuvel LP. The impact of COVID-19 pandemic on the diagnosis and management of inborn errors of metabolism: A global perspective. Mol Genet Metab. 2020 Nov;131(3):285-288. doi: 10.1016/j.ymgme.2020.09.004. Epub 2020 Sep 25. PMID: 33004274; PMCID: PMC7518833.
7. Brunetti-Pierri N, Fecarotta S, Staiano A, Strisciuglio P, Parenti G. Ensuring continuity of care for children with inherited metabolic diseases at the time of COVID-19: the experience of a metabolic unit in Italy. Genet Med. 2020 Jul;22(7):1178-1180. doi: 10.1038/s41436-020-0831-4. Epub 2020 May 15. PMID: 32409735.
8. Xiong J, Lipsitz O, Nasri F, Lui LMW, Gill H, Phan L, Chen-Li D, Iacobucci M, Ho R, Majeed A, McIntyre RS. Impact of COVID-19 pandemic on mental health in the general population: A systematic review. J Affect Disord. 2020 Dec 1;277:55-64. doi: 10.1016/j.jad.2020.08.001. Epub 2020 Aug 8. PMID: 32799105; PMCID: PMC7413844.

How to cite this article: Gugelmo G, Schiff S, Lovato E, Lenzini L, Boscari F, Francini-Pesenti F, Avogaro A, Vitturi N. Lifestyle, Nutritional and Psychological Changes in Adult Patients with Inborn Errors of Metabolism during COVID 19 Pandemic: Results from an Online Survey. J Biomed Res Environ Sci. 2021 Mar 02; 2(3): 100-103. doi: 10.37871/jbres1197, Article ID: JBRES1197